

# Sporadic Nocturnal Frontal Lobe Epilepsy--Report on Two Cases and Review of the First Taiwanese Series of 10 Cases

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## Abstract-

**Purpose:** To report two additional cases of sporadic (i.e. non-familial) Nocturnal Frontal Lobe Epilepsy (NFLE) and integrate these two cases within the first series of 10 cases of sporadic NFLE reported in Taiwanese patients, and compare the findings with familial NFLE and with findings from Caucasian NFLE patients.

**Methods:** Clinical interviews, neurological examinations, EEG, brain MRI, and overnight video-polysomnographic (vPSG) monitoring with EEG seizure montage, and treatment outcome.

**Results:** The two additional patients were 12 and 29 year old females manifesting their sporadic NFLE with paroxysmal arousals (PAs) and nocturnal paroxysmal dystonia (NPD), respectively, and also hypermotor seizure behavior in one of these patients. In the series of 10 Taiwanese cases, 3 were classified with PAs, and 7 with NPD. No patient had combined PA/NPD seizure types. Furthermore, 4 cases also demonstrated hypermotor seizure behavior. Gender ratio was four males to six females. Mean age of NFLE onset was 9.6 yrs (range, 1-23), mean age at initial presentation was 16.1 yrs (range, 2-41), and mean age at latest follow-up was 23.1 yrs (range 11-45). Premorbid history was negative for any neurologic, medical or psychiatric disorder. MRI brain scan abnormalities with clinical correlates were found in two patient. During vPSG studies, four of ten patients with NFLE seizure events had concurrent epileptiform EEG activity, and two patients had interictal epileptiform EEG activity during their vPSG studies. No case had a spontaneous remission. Anticonvulsant therapy was highly effective in all ten cases (>75% reduction in seizure frequency).

**Conclusion:** The two newly reported cases that were integrated into the first series of 10 Taiwanese patients with sporadic NFLE corresponds closely to previously reported sporadic and familial NFLE among Caucasian patients in Europe and North America. There was a high rate of sustained anticonvulsant treatment efficacy, particularly with carbamazepine, oxcarbamazepine, and topiramate. Also, 4 of the 10 patients had hypermotor manifestations (in part) of their NFLE (including one of the two newly reported cases), which are discussed in regards to the newly published entity of "Sleep-Related Hypermotor Epilepsy(1)."

**Key Words:** Nocturnal frontal lobe epilepsy, Nocturnal paroxysmal dystonia, Paroxysmal arousals, Sleep-Related Hypermotor Epilepsy, Anticonvulsant therapy

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## INTRODUCTION

A distinct form of clear-cut seizures arising from epileptic foci located within the frontal lobe (in particular in mesial and orbital cortices) and emerging almost exclusively from sleep has been described with the term of nocturnal frontal lobe epilepsy (NFLE) since the 1990s<sup>(2-5)</sup>. The manifestations of NFLE<sup>(6)</sup> are characterized by a wide spectrum of clinical features: assumption of postures, rhythmic and repetitive movements of arms and legs, rapid uncoordinated movements, with dystonic or dyskinesic components, complex motor activities (ambulation, wandering, pelvic thrusting), sudden elevation of the trunk and head associated with expression of fear and vocalization.

On the basis of the different intensity, duration and features of the motor patterns, Provini et al.<sup>(5)</sup> classified the nocturnal FLE (NFLE) epileptic seizures into three groups, according to Montagna<sup>(7)</sup>: (1) Paroxysmal Arousals (PA) with brief (< 20 sec) episodes in which patients suddenly open their eyes, raise their heads or sit up in bed with a bizarre posture of the limbs, staring around with a frightened or surprised expression, and sometimes screaming; they then return to sleep. (2) Nocturnal Paroxysmal Dystonia (NPD) with a longer duration (20sec–2min) and more complex behaviors characterized by wide-ranging, often violent, and sometimes ballistic movements, with dystonic posturing of the head, trunk and limbs, such as head rotation, torsion of the trunk and choreo-athetoid movements of the arms and legs, with vocalization. (3) Episodic Nocturnal Wandering(ENW) with duration of episodes lasting up to 1–3min, for which the characteristic feature is stereotypic paroxysmal ambulation during sleep, often with agitation and accompanied by screaming and bizarre, dystonic movements.

Sporadic (i.e.non-familial) NFLE is rarely reported, in contrast to familial NFLE, and therefore is poorly understood. We have recently reported on a series of eight consecutive cases of sporadic NFLE that comprised the first case series in Taiwan, or any Asian country,<sup>(8)</sup> and now we wish to present two additional cases to expand the spectrum of knowledge of this under-recognized sleep-related disorder, and to summarize the findings in the updated series of 10 cases. We will provide clinical and

vPSG findings, particularly vis-à-vis findings reported in Caucasian populations. The differential diagnosis will be presented. Furthermore, our findings will be discussed in the context of the newly recognized diagnostic entity of "Sleep-Related Hypermotor Epilepsy"<sup>(1)</sup>.

## METHODS

The two additional cases, Patients 2 and 10 listed in Table 1 (12 year old and 29 year old females, with case summaries presented below) were part of a series of ten consecutive patients (6 female and 4 male), who had presented to the sleep clinic of one author (S-BY) from July 2006 to July 2015 on account of nocturnal paroxysmal episodes suggestive of NFLE. Mean age of NFLE onset was 9.6 yrs (range, 1-23), mean age at initial presentation was 16.1 yrs (range, 2-41), and mean age at latest follow-up was 23.1 yrs (range 11-45). These ten patients completed a comprehensive questionnaire covering life time sleep-wake, medical and psychiatric history, and review of systems. The patients and, when applicable, their caregivers were interviewed. They also received a full neurological examination by a pediatric or adult neurologist. Routine daytime awake and sleep EEG recordings were also performed for these patients. FLEP (Frontal Lobe Epilepsy and Parasomnias) scale scores<sup>(9)</sup> were performed for each patient. Table 1 contains the clinical data, including results of brain MRIs. The final column in Table 1 contains the FLEP scores for the ten patients.

An overnight, hospital-based, vPSG monitoring, utilizing standard recording and scoring methods<sup>(10)</sup>, was then performed on these patients after discontinuation of anti-epileptic drugs (AED) for at least one day, except patient 9 who had recurrent attacks several times daily in wakefulness and sleep beginning shortly on the day of medication discontinuation. The PSG monitoring included an electrooculogram (EOG), expanded EEG (seizure montage) with a 1cm/sec recording speed, submental and bilateral anterior tibialis electromyograms (EMGs), nasal-oral airflow, chest and abdomen respiratory effort, electrocardiogram, and continuous time-synchronized audiovisual recording.

We analysed all the video-polysomnographic recordings and summarized all the pathological motor

**Table 1.** Clinical data, total 10 patients

Patient	Sex	Age of latest visit, yr	Age of initial visit, yr	Age of seizure onset, yr	Duration of seizure history (yrs)	Trigger factor	Personal antecedents	Seizure type	Neurological image (MRI)	Positive response to antiepileptic drugs	FLEP score (+ numbers)
1	M	19	9	5	14	Negative	negative	NPD	unremarkable	Yes, oxcarbazepine 600mg bid and topiramate 25mg hs	6
2	F	22	12	12	10	Negative	negative	PA	right temporal vasculature lesion (fig1)	Yes, carbamazepine 200mg bid and topiramate 50 mg bid	4
3	M	18	8	2	16	Protacted exercise	negative	NPD	unremarkable	Yes, carbamazepine 200mg qd & 300 mg hs and topiramate 50 mg	4
4	M	11	2	1	10	Negative	negative	NPD	unremarkable	Yes, carbamazepine 200mg hs	5
5	F	16	8	8	8	Negative	negative	NPD	right orbitofrontal lobe cortical	Yes, oxcarbazepine 300mg bid and topiramate 25mg bid	6
6	F	45	41	23	22	Upper respiratory viral infections	negative	PA	unremarkable	Yes, carbamazepine 400mg bid	4
7	M	11	7	5	6	Negative	negative	PA	unremarkable	Yes, valproate 300mg bid and levetiracetam 500 mg bid	3
8	F	37	30	20	17	Menstrual related	negative	NPD	unremarkable	Yes, carbamazepine 200mg bid or lamotrigine 300mg hs	4
9	F	22	15	6	16	Negative	negative	NPD	unremarkable	Yes, acetazolamide 250mg bid, oxcarbazepine 300mg hs, and clonazepam 0.5mg tid	7
10	M	30	29	14	16	Negative	negative	NPD	unremarkable	Yes, oxcarbazepine 300 mg hs, topiramate 100 mg hs and acetazolamide 250mg hs	4

events, as contained in table 2. We examined the EEG tracing and compared the seizures semeiology with the EEG patterns. All ten cases had the diagnosis of NFLE according to literature data<sup>(5)</sup>, when patients displayed one or more motor episodes, of any kind of intensity and duration, associated with clear-cut ictal epileptiform activity during polysomnography. When ictal EEG was uninformative, we required the recording of two or more seizures with a stereotypic motor pattern. In spite of patient 3 having only one episode attack during the vPSG, the typical presentation of choreo-dystonic movements during sleep, together with a beneficial response to AED, allowed for the definite diagnosis of NFLE in this patient.

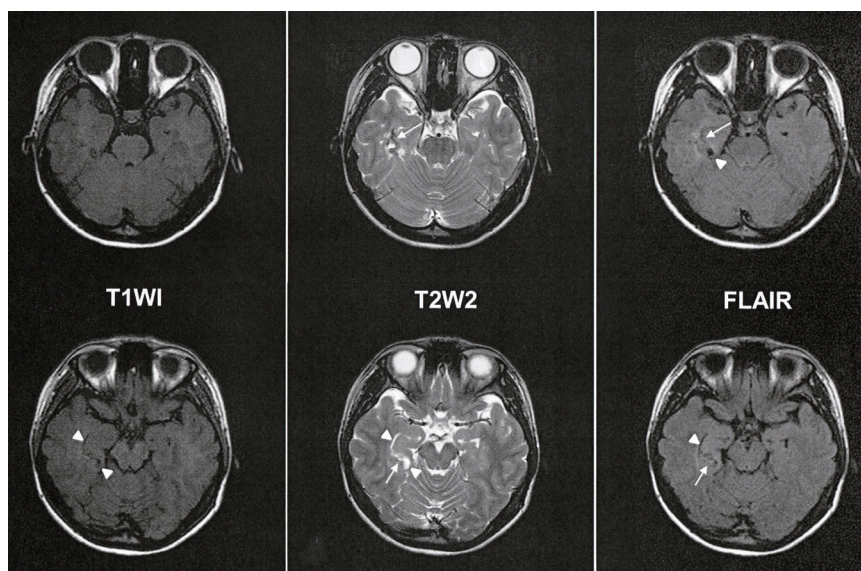
## RESULTS

Case Summaries will now be presented for the two newly reported patients (Cases 2, 10): Case 2: A 12 year old girl presented with her parents who reported paroxysmal eye-opening with fixed staring during nocturnal sleep, accompanied by lip movements as if she were talking. There were also bizarre upper limb

movements. During the preceding 6 months, these attacks occurred nearly every night, and occasionally there were several episodes per night. Most episodes lasted for several minutes (as told by her mother), and the girl was always amnesic for the episodes.

Video- PSG results: there was one episode of sudden onset eye-opening with staring and upper limb dystonic posturing emerging from N2 sleep, associated with epileptiform activity.

Case 10: A 29 year-old female nurse presented with the chief complaint of paroxysmal bizarre movements with moaning during nocturnal sleep since the age of 14 years. Her bizarre movements were noted by her roommate in a school dormitory, with attacks occurring nearly every night, and occasionally there were several episodes per night. The duration of the attacks lasted for several minutes, according to the roommate. She was always amnesic for the episodes. She had consulted with an epilepsy specialist, and was told she had a seizure disorder, although an awake and sleep EEG and brain MRI were all unremarkable. She then tried many AEDs, with little benefit. She visited various other neurologists for help,



**Figure 1.** Brain MRI of Patient 2 reveals right medial temporal lobe flow void tubular structure (white arrows) with multicentric encephalomalacia (arrow heads) and surrounding brain parenchymal gliosis-like change, possibly vascular lesion such as arteriovenous malformation or arteriovenous fistula with old brain insult.

but the nocturnal attacks persisted unabated. A community neurologist then referred the patient to the sleep clinic of one of the authors (S-B Y) for further sleep evaluation and sleep lab study, as the neurologist suspected the diagnosis of nocturnal seizures despite all the normal findings from his repeated examinations and testing.

Video-PSG results: there were four episodes of sudden-onset raising of the head with dystonic-chorea movements from stage N1 sleep (1 episode) and from stage N2 sleep (3 episodes), there were not associated with epileptiform activity.

For the entire series of 10 consecutive patients, 9 of 10 patients manifested one or more nocturnal attacks during vPSG monitoring. Patient 9 (who was maintained on anticonvulsant medication) did not have an attack, but she had interictal epileptiform discharges (spike and waves) during the overnight vPSG study, and attacks were viewed with event video home recording provided by her family. These ten cases were classified as PA (three cases) and NPD (seven cases). All ten cases had sporadic NFLE, without any positive family history. No patient had combined PA/NPD seizure types.

The nocturnal paroxysmal episodes had been present for up to 22 years (mean 13.5 yrs) before the current

reported evaluation; age at presentation ranged from 11 to 45 yrs (mean 23.1 yrs). Mean duration of the seizure history was 13.5 yrs (range, 6–22 yrs). Seizure frequency in all ten patients was several attacks nightly, during nearly every night. All ten patients were unaware of their nocturnal motor manifestations, and so medical consultation was sought by their families who had observed the recurrent episodes. All ten patients had undergone neuroradiological examination, viz. brain MRI. Abnormalities with clinical correlates were detected in two cases; one involved a right temporal lesion that was a suspected vascular lesion in patient 2 (Fig. 1), and the other had a right orbitofrontal lobe cortical dysplasia in patient 5<sup>(11)</sup>. Three patients (patient 3, 5, and 9) also had occasional seizures during daytime wakefulness, similar to their seizures during sleep. In these three cases, however, daytime seizures were sporadic with low frequency. Only patient 2 and 10 had rarely secondarily generalized seizure following the event episode.

Seven PA episodes were recorded from three patients, lasting a mean of 19.0 sec in patient 2, 11.0 sec in patient 6 and 11.5 sec in patient 7. In the PA episodes, the first movement usually involved the upper limbs: the patients suddenly raised their arms while asleep, or assumed a

**Table 2.** Videopolysomnographic data, total 10 patients

Patient	Seizure type	Number of seizure recorded	Seizure duration (seconds, mean±SD)	First seizure onset after sleep onset (minutes)	Sleep stages during which seizure appeared				Autonomic modification	Seizure symptom presentation
					stage1	stage2	stageSWS	stage REM		
1	NPD	12	41.0±4.9	22	1	9	1	1	Yes, tachypnea and tachycardia	Dystonic posture, arms/legs; crying; hypermotor behavior of legs, tachypnea
2	PA	1	19	338	0	1	0	0	Yes, tachycardia	Eyes staring, moaning, dystonic posture, arms/legs
3	NPD	1	34	278	0	1	0	0	No	choreo-dystonic movement
4	NPD	6	30.5±12.3	64	0	5	1	0	No	crying with dystonic posture, followed by hypermotor behavior of legs
5	NPD	6	35.8±7.5	3	0	6	0	0	Yes, tachycardia	Dystonic posture, arms; screaming with terrified expression
6	PA	4	11.0±0.8	223	1	2	0	1	Yes, tachycardia	Dystonic posture, legs; moaning
7	PA	2	11.5±0.7	162	0	2	0	0	No	right upper limb elevation with dystonic posture
8	NPD	3	35.3±6.7	299	0	3	0	0	Yes, tachycardia	turn to prone position; dystonic posturing; screaming with hypermotor behavior
9	NPD	0	N/A	N/A	0	0	0	0	N/A	N/A, but dystonic posture by family video recording
10	NPD	4	46.3±14.9	47	1	3	0	0	No	raising head, followed by choreo-dystonic movements and hypermotor

**Table 3.** Interictal and ictal EEG activity, total 10 patients

Patient	Seizure type	Interictal routine daytime awake EEG abnormality	Interictal routine daytime sleep EEG abnormality	Interictal overnight polysomnographic EEG abnormality	Ictal overnight polysomnographic EEG abnormality
1	NPD	negative	negative	negative	burst sharp waves
2	PA	negative	negative	negative	burst sharp waves
3	NPD	negative	negative	negative	negative
4	NPD	negative	negative	burst sharp waves	negative
5	NPD	negative	repetitive spike/waves over F4-C4 and F3-C3	negative	background EEG activity suppression and following the burst sharp waves
6	PA	negative	negative	negative	continuous rhythmic sharp activity (2-6 Hz) for about 50 seconds and following by the slow waves(1-2 Hz)
7	PA	negative	negative	negative	negative
8	NPD	negative	negative	negative	negative
9	NPD	negative	focal spike over right frontal region	runs of spike and waves	N/A
10	NPD	negative	negative	negative	negative

dystonic posture of one hand. Thirty-two NPD episodes were recorded from seven patients, lasting from 22 to 65 sec (mean 46.3±14.9 sec). There was a mean 4.6 NPD

episodes per vPSG recording. Seizures appeared between 3 and 338min after sleep onset (mean 159.6 min). 94.9% (37/39) of the episode attacks appeared during NREM

sleep, with 89.7% of the attacks during light (stage N1–N2), and 5.1% during deep sleep (stage N3). Only 5.1% of seizures occurred during REM sleep. Mean seizure duration in the six NPD patients ranged from 30.5 sec to 46.0 sec, and mean seizure duration in the three PA patients was 11.0 to 19.0 sec.

Table 3 showed the results of routine daytime awake and sleep EEG study and the overnight polysomnographic EEG presentation. All ten cases had normal wake scalp EEG activity. In eight patients (80%), sleep EEG was completely normal. Only two patients (20%) (patient 5 and 9) showed focal frontal epileptic abnormalities. EEG recordings during the attacks failed to disclose ictal epileptic activity in five cases. In such cases, EEG was often masked by muscle artifacts or characterized by an abrupt transition to wake activity or light sleep, occasionally (patient 7) preceded by a K-complex. EEG recordings during the attacks did record ictal activity in four cases: a burst of sharp waves in patient 1 and patient 2; background EEG activity suppression followed by a burst of sharp waves in patient 5<sup>(11)</sup>; and continuous rhythmic sharp activity (2–6 Hz) for 50 sec, followed by slow-wave activity (1–2 Hz) in patient 6. The first EEG modifications were either a diffuse flattening of background activity (1 case), or sharp activity (three cases). In one patient (patient 9), the interictal EEG was characterized by spike-and-waves activity. Autonomic hyperactivity was remarkable in many cases (Table 2): tachycardia (four cases) and sustained tachypnea with tachycardia (one case) appeared synchronously with seizure onset or accompanied the movement artifacts.

The FLEP scores for the ten patients were all more than 3 scores (mean:  $4.7 \pm 1.3$ ), strongly supported of NFLE. FLEP (Frontal Lobe Epilepsy and Parasomnias) scale scores were calculated for each patient, according to the method of Derry et al.<sup>(9,12)</sup>. A FLEP score  $\geq 1$  strongly supports the diagnosis of NFLE and FLEP score of  $\leq 0$  strongly supports the diagnosis of a parasomnia (viz. NREM sleep arousal parasomnia, such as sleepwalking, sleep terrors, confusional arousals).

Based on prior reported experience<sup>(13,14)</sup>, carbamazepine was the drug of first choice in these patients. Due to safety considerations<sup>(15)</sup>, oxcarbamazepine was also the initial treatment of choice. Five patients received carbamazepine at a dosage varying from 200 to 800mg/

day in monotherapy (two cases) or polytherapy (three cases, combined with topiramate or lamotrigine). Four patients received oxcarbamazepine at a dosage varying from 300 to 1200 mg/day in polytherapy (two combined with topiramate, one is combined with topiramate and acetazolamide, and the last one combined with acetazolamide and clonazepam). Patient 7 was referred from a pediatric neurologist and was still under control with sodium valproate therapy, 300 mg bid and levetiracetam, 500 mg bid. These medications reduced the frequency of nocturnal seizures by at least 75% and abolished any occasional diurnal attack by more than 90%. None of the patients had any adverse effects or complaints related to treatment.

## DISCUSSION

Our expanded case series of 10 sporadic (non-familial) NFLE, including the addition of 2 additional cases, which comprise the first case series reported in Taiwan, or from any other Asian country, corresponds closely to previously reported sporadic and familial NFLE cases among Caucasian patients in Europe and North America.

Our findings reinforce how NFLE should always be suspected in the presence of paroxysmal nocturnal motor events characterized by a high frequency of same-night or inter-night recurrence, persistence beyond puberty into adulthood, quasi-extrapyramidal features, agitated behavior and remarkable stereotypy of the attacks.

Although EEG recordings during the attacks failed to disclose ictal epileptic activity in five cases, and four cases also had no interictal findings, we concluded that the events were seizures because of the stereotyped movements and behaviors, and also because of the robust response to anti-epileptic medications. The absence of scalp EEG ictal epileptic activity during the NFLE seizure attacks in some patients can be explained by the seizure focus being located in deep brain regions, as previously discussed<sup>(16)</sup>. In these cases of the absence of scalp EEG ictal epileptic activity during the NFLE seizure attacks, only sphenoidal and intracerebral EEG recordings can help to identify ictal and/or interictal discharges<sup>(2,16)</sup>.

Mai et al.<sup>(17)</sup> reported the electro-clinical analysis of the ictal episodes in some cases, suggested an extra-frontal origin of the attacks, with the term of NFLE are likely

included some patients with complex nocturnal motor automatisms of extra-frontal origin. This could explain our patient 2 in this case series, in spite of her right temporal lesion, the patient still present the manifestation NFLE due to abundant connection between frontal and temporal lobe.

The high rate of sustained treatment efficacy is gratifying for the patients (once they are informed of their disorder), their families, and their physicians. The most commonly effective anticonvulsants, as first-line therapy, in our series were carbamazepine and oxcarbazepine, but also topiramate was effective in five patients, and several other anticonvulsants were used with benefit. Topiramate was reported to be highly effective therapy in a series of 24 consecutive NFLE patients presenting to a sleep disorders center in Italy, with 15 cases being sporadic NFLE and 9 cases being familial NFLE<sup>(6)</sup>. Nearly 90% of topiramate-treated patients (as monotherapy or add-on therapy) became either seizure-free or had >50% reduction in seizure frequency. Finally, we needed to mention the treatment experience of patient 10, the female patient was diagnosed as sleep-related epilepsy since 17 y/o, and tried various anti-epilepsy medication before visiting our sleep clinic, such as phenytoin, valproate, carbamazepine, oxcarbazepine, topiramate, levetiracetam, vigabatrin, lamotrigine, zonisamide, and so on. But these AED usually improved less than 50%. Varadkar et al.<sup>(18)</sup> reported three members of a family ADNFLFLE with intractable control that responded to acetazolamide (ACZ) as add-on therapy to carbamazepine (CBZ). According to past treatment and Varadkar et al. experience, we selected carbamazepine, topiramate and acetazolamide before bedtime use for this patient, her nocturnal episode attacks dramatically improved more than 90% till now. As ADNFLFLE is suspected to be a channelopathy, Varadkar et al. hypothesized that ACZ may be exerting its therapeutic effect through this action, rather than by potentiating CBZ.

Four of our 10 patients (#1, 4, 8, 10) in our series had hypermotor presentations with their sporadic NFLE. Recently, the clinical entity of "Sleep-Related Hypermotor Epilepsy" (SHE) has been published, which was based on a consensus conference of international experts who gathered in Bologna, Italy in September 2014<sup>(1)</sup>. The rationale for this consensus conference and the generation of the definition and diagnostic criteria of SHE was based on the "evidence that the attacks are associated

with sleep rather than time of day, the seizures may arise from extrafrontal sites [i.e. temporal lobe origin], and the motor aspects of the seizures are characteristic." Three levels of diagnostic certainty were developed: witnessed seizures (possible) SHE, video-documented (clinical) SHE, and video-EEG-documented (confirmed) SHE. The core clinical features of SHE involve the brief (<2 min) duration of seizures with stereotyped motor patterns within individuals, and abrupt onset and abrupt offset. The most common clinical expression consists of "hypermotor" events, although a spectrum of symptoms ranging from PA to NPD to hypermotor events do occur, as demonstrated in the case series of 10 patients reported herein. It should be emphasized that SHE includes the spectrum of PA, NPD, and ENW, and is the updated, current term for NFLE. Although most SHE seizures occur from sleep, sometimes they can occur from wakefulness. This important new classification of SHE is relevant to the differential diagnosis to now be discussed, in regards to the validity in comparing NFLE from nocturnal temporal lobe epilepsy.

Sleep-related hyperkinetic seizures of temporal lobe origin were documented in three patients by means of long-term stereo-EEG investigations and surgical outcome<sup>(19)</sup>. In a retrospective study of 442 consecutive patients with drug-resistant hyperkinetic seizures that were surgically treated, 25 of these patients had sleep-related hyperkinetic seizures, of which 18 had a frontal lobe onset, and 7 had a temporal lobe onset<sup>(17)</sup>. The latter group of seven patients with temporal lobe origin had anamnestic and clinical features that mirrored the clinical features found in the 18 patients with frontal onset, with agitated movements, high seizure frequency and absent history of febrile convulsions. Therefore, the presence of sleep-related hyperkinetic seizures is not specific to NFLE, as they can also be found with nocturnal temporal lobe epilepsy, which was a major point of emphasis in the recent publication on SHE<sup>(1)</sup>.

In regards to the differential diagnosis of NFLE with parasomnias, clues for distinguishing nocturnal seizures from sleep terrors include the following<sup>(20)</sup>: (i) nocturnal seizures have a brief, stereo-typical presentation; (ii) with frontal lobe seizures, events usually occur from sleep (NFLE), with bizarre hypermotor activity, but without altered consciousness upon awakening; (iii) sleep terrors

rarely occur >1 time per night, and very rarely occur nightly, and almost never occur nightly for months or years; (iv) with sleep terrors, there is confusion and disorientation during an arousal, and episodes can last for minutes; (v) sleep terrors usually respond to bedtime benzodiazepine therapy. The topic of distinguishing nocturnal seizures from parasomnias has recently been reviewed and discussed<sup>(21)</sup>.

In conclusion, differentiating NFLE from nocturnal temporal lobe epilepsy, NREM sleep parasomnias, and other nocturnal conditions, requires a careful, comprehensive, and multi-modal clinical evaluation, given the peculiar (and even bizarre) presentation of the recurrent abnormal behaviors that often emerge in the context of a normal scalp EEG. The newly recognized entity of SHE<sup>(1)</sup> allows for the inclusion of both frontal lobe and temporal lobe origins of recurrent sleep-related hyperkinetic seizures, and provides a unifying diagnosis.

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